Choanal Atresia: Surgical Management by Hegar's Dilators

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Abstract Choanal atresia is one of the more commonly observed congenital abnormalities of the nose. The condition is predominantly found in females with a M:F ratio of 1:2. 65–75 % of patients with choanal atresia are unilateral, and the rest are bilateral. About 50 % of the cases are associated with other congenital anomalies, the most common being coloboma, Heart disease, choanal atresia, mental and growth retardation, genital hypoplasia, ear deformities syndrome. The embryonic origin of choanal atresia is due to persistent bucco-pharyngeal or naso-buccal membrane. Our study was a retrospective study of 14 cases of choanal atresia. All the cases were operated by the first author. Each patient was investigated in detail and recorded by stratifying in sex of patient, age of presentation, type and site of atresia, and associated other congenital anomalies. Patient were operated under general anesthesia and Hegar's dilators were used to perforate the atretic plate as Hegar's dilators are believed to have ideal curvature with respect to the sloping contour of the nasal floor. Out of the total 14 cases, 9 (64 %, $n = 14$) cases were male and 5 (36 %, $n = 14$) females. The age variations varied from youngest of 3 days to the oldest 31 year old female. Stents were kept for a period of 6–8 weeks and regular follow up nasal endoscopy was done weekly for suctioning and visualizing the size of the airway. There are five different surgical approaches that have been described for surgical treatment of choanal atresia: (1) trans-nasal, (2) trans-palatal, (3) trans-septal, (4) trans-antral and (5) sublabial–transnasal. In our study M:F ratio was 1.8:1 which is comparable Gosepath et al. (Rhinology 45:158–163,

2007) (2:1). In our study bilateral atresia was seen in 43 % (6, $n = 14$) and unilateral in 57 % (8, $n = 14$) which is comparable to Newman et al. (44 % bilateral cases, $n = 43$). Amicable and prompt referral to the ENT surgeon can be of immense value so as to buy adequate time for the surgeon to decide and act as per the patient's clinical and radiological findings. Hegar's dilator is passed along the floor of the nose, pushed against the septum so as to avoid penetrating the basal sphenoid.

Keywords Choanal atresia · Hegar's dilator · Pediatric · Choana

Choanal atresia is one of the more commonly observed congenital abnormalities of the nose [1]. This condition was first described by Johann George Roederer in 1755. This was later termed as an anatomical abnormality of palatine bone by Adolf Otto in 1854 [2], Carl Emmert has been successfully credited with the first choanal atresia repair in 1854. The word “Choana” is derived from the Greek word Xovan, meaning funnel hence the term “Posterior Choana” would literally mean a posterior funnel [1]. Incidence of choanal atresia is around 1 in 7,000 live births [3]. It is a potential life threatening condition in neonates because this condition is predominantly in first 6 weeks of births [4]. The condition is predominantly found in females with a M:F ratio of 1:2 [5]. 65–75 % of patients with choanal atresia are unilateral, and the rest are bilateral [6]. 30 % of cases are pure bony, whereas 70 % are mixed bony-membranous [7]. About 50 % of the cases are associated with other congenital anomalies, the most common being Coloboma, Heart disease, Choanal atresia, Mental and growth retardation, Genital hypoplasia, Ear deformities (CHARGE) syndrome [8].

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Embryology

The development of choanae takes place between the 4th and 11th weeks of gestation [9]. The embryonic origin of choanal atresia is due to persistent bucco-pharyngeal or naso-buccal membrane [10].

Pathophysiology

The infant are obligate nasal breather, and they breathe through mouth only during crying and reduction in the diameter of the nasal airway increase nasal airway resistance and hence result in respiratory distress in neonates. Crying instantly relieves the airway obstruction. About 50 % of patients with choanal atresia have the CHARGE syndrome. If patency of a neonate's airway is dubious a catheter should be passed through each nostril and into the child's oral cavity.

Materials and Methods

Our study was a retrospective study of 14 cases of choanal atresia. All the cases were operated by the first author. Each patient was investigated in detail and recorded by stratifying in sex of patient, age of presentation, type and site of atresia, and associated other congenital anomalies. The diagnosis was made by clinical presentation of neonates and then in suspected cases by the inability to pass a small suction catheter through the nares and confirmed by CT scans and nasal endoscopy. All patients were operated for surgical repair of choanal atresia through a transnasal technique. A 4 mm, 0° nasal endoscope was used for visualization before and after the dilatation. Curved Hegar's dilator was used to perforate and then subsequently dilate the atretic plate transnasally. After adequate opening was achieved stenting using portex endotracheal tubes was done in all the cases using 'Vikas Sinha Stenting' procedure and stent was kept for a period 7–15 days as per the tolerability of the patients. All patients were given post-operative prophylactic antibiotics and were kept in Intensive care unit setup for a period of 48–72 h post operatively. Subsequent serial dilatation was performed in a few cases after 6 weeks as per the postoperative nasal endoscopic and CT scan finding.

Surgical Technique

Patient were operated under general anesthesia and Hegar's dilators were used to perforate the atretic plate as Hegar's

dilators have ideal curvature with respect to the sloping contour of the nasal floor. The atretic plate is thinnest and weakest at the junction of the floor and the posterior end of the septum and can be easily perforated using smallest Hegar's dilator to avoid trauma to surrounding tissue. Hegar's dilator is passed along the floor of the nose, pushed against the septum so as to avoid penetrating the basal sphenoid. The index finger of the left hand was positioned at the nasopharynx against the nasopharyngeal side of the atretic plate with the aim of safeguarding against accidental sudden perforation and preventing the accidental trauma to the base of the skull. Gentle uniform pressure and screwing movement was applied using the Hegar's dilator till the atretic plate was perforated then it was serially punctured and dilated using increasing sizes of Hegar's dilator. The maximum size of the dilator was approximately equal to size of external nares of the child. The procedure was done bilaterally till adequately dilated choana were achieved on both sides. In few cases Bone Rongeur was used to nibble the atretic plate after the initial perforation by Hegar's dilator.

Stenting Procedure

Vikas Sinha stenting procedure was used in all the cases. A noncuffed portex endotracheal tube was used. The size of the tube was approximately the size of the last Hegar's dilator which was used without any apparent resistance. The tube was folded at the midpoint thus making two exactly equal lengths then posterior fenestration (outer curved side) of the tube at the midpoint was created with a number 11 blade which allows air and nasal secretions to pass from the nose to the nasopharynx while the anterior part of the tube (inner curved end) remains in continuity [1] thereafter smaller vents or holes were made in a different site and axis for the drainage of nasal secretions. Following the preceding procedure two red rubber catheters were passed from both nostrils to be taken out from the oral cavity then two ends of the newly created stent were tied with a red rubber catheter and pulled retrograde from the nasal cavity simultaneously so that posterior fenestration lies exactly in the midline at the nasopharynx. An equal length of the two tubes outside of the nose will confirm that the posterior fenestration is in midline and in the nasopharynx. Suction clearance was done through the stent to prevent blockage from blood clot and secretions. The tube was secured at the nostril with along with a small gauze piece at the columella to prevent damage to columella as continual friction of silk suture against the columella causes pressure necrosis on the columella. All the postoperative patients were kept in the intensive care unit for regular suction of the nasal passage for 1 week.

Results

Out of the total 14 cases, 9 (64 %, $n = 14$) cases were male and 5 (36 %, $n = 14$) females. The age variations varied from youngest of 3 days to the oldest 31 year old female. 6 (43 %, $n = 14$) patients had bilateral choanal atresia while 4 (28.5 %, $n = 14$) cases had right side unilateral and 4 (28.5 %, $n = 14$) cases left side unilateral choanal atresia. Most commonly diagnostic maneuver used to establish the diagnosis was the inability to pass a rubber catheter through the nose into the nasopharynx. Nasal endoscopy was performed in all cases with 0° endoscope. CT scan were performed in all cases which depicted five cases as bony and nine cases were of mixed variety i.e. both bony and membranous and the finding were further confirmed peroperatively. All the cases were operated endonasally along with perforation of atretic plate using Hegar's dilators. The success criteria of the surgery was passing of the stent successfully through both the nasal cavity. In all the cases adequate airway was established on the table by initial perforation then subsequent dilatation using increasing sized Hegar's dilators. No life threatening intraoperative complication like skull base injuries or spinal injuries were observed in any of the cases operated. In all the patients, subsequent stenting was done using portex non cuffed endotracheal tube by 'Vikas sinha stenting technique', the size of the tube was approximately the size of the last Hegar's dilator used while dilatation. Five cases (36 %, $n = 14$) of neonates were associated with CHARGE syndrome and all failed to survive for more than 5–10 days due to the complications of CHARGE syndrome and unrelated to the outcome of surgery. Stents were kept for a period of 6–8 weeks and regular follow up nasal endoscopy was done weekly for suctioning and visualizing the size of the airway. Normal saline drops were used to avoid crusting and hence blockage of the airway. Five patients (36 %, $n = 14$) required repeated dilatation after 6 weeks of dilatation and stenting. Columellar necrosis developed in two cases (14 %, $n = 14$) which compelled us to remove the stent around 3 weeks after the dilatation in both the cases. All the patients were regularly followed up for a period of 12–24 months. In two patients, granulations developed around the stent. The stent was removed subsequently and Foleys catheter was used as stent where the balloon part of catheter was insinuated at choana like a dumbbell, where the balloon part of catheter was on either side of choana.

Discussion

Choanal atresia is a relatively rare but possibly fatal disorder. Maintenance of airway is of prime importance

especially in bilateral cases. There are five different surgical approaches that have been described for surgical treatment of choanal atresia: (1) trans-nasal, (2) trans-palatal, (3) trans-septal, (4) trans-antral and (5) sublabial-transnasal [11, 12]. The most popular are trans-palatal and trans-nasal approaches. Hegar's dilator was used in all our patients since Hegar dilator's curve attunes with the natural curve of the palate and the floor of nose and its blunt curvatures has less tendency of damaging surrounding structures. In our study M:F ratio was 1.8:1 which is comparable Gosepath et al. [13] (2:1). In our study bilateral atresia was seen in 43 % (6, $n = 14$) and unilateral in 57 % (8, $n = 14$) which is comparable to Newman et al. [14] (44 % bilateral cases, $n = 43$). Portex endotracheal uncuffed tube was used as stent whose size was dependent on the size of last Hegar's dilator which could be introduced without resistance as done by Gujarathi et al. [15] and Sharma et al. [16]. Nasal stents were used for a period of 6 weeks which is comparable to Al-Amman [17] and earlier study of Sinha et al. [1]. Use of stent is controversial [18] as stated in literature because it may cause discomfort, localized infection and ulceration as in our study 2 (14 %, $n = 14$) developed columellar necrosis. Five cases (36 %, $n = 14$) cases required revision dilatation even after 6 weeks of stenting, variable results have been reported in literature i.e. study of Uzomefuna et al. [19] (57 % in cases with stent and 33 % in cases without stent) and Velegrakis et al. [20] (57 % in bilateral and 25 % in unilateral).

Conclusion

"Birth without breath" is the precise description of choanal atresia. It is a rare disorder in routine practice but can be life threatening if not diagnosed accurately and timely. Both the pediatrician and ENT surgeon should be oriented to the presenting symptoms and the anticipated clinical picture in such cases. Amicable and prompt referral to the ENT surgeon can be of immense value so as to buy adequate time for the surgeon to decide and act as per the patient's clinical and radiological findings. The surgery for choanal atresia should be undertaken as soon as the diagnosis is made. Intranasal Hegar's dilator method is a very safe method to perforate the atretic plate whether bony or membranous. Controlled pressure of the Hegar's dilator using a twisting movement is the key for the successful controlled perforation of the atretic plate. Stenting with either an endotracheal tube ("Vikas Sinha Stenting") or balloon is mandatory to prevent re stenosis. Repeated serial dilatation may be performed in cases where early signs of re stenosis can be visualized on CT scan and nasal endoscopy. Various approaches have so far been used in treatment of choanal atresia and variable results have been

reported in the literature but it will not be wrong to highlight the fact that without prompt diagnosis at the primary level or by the pediatrician it can prove as a potentially fatal condition, hence early diagnosis, meticulous evaluation and expeditious and rational surgical management are the essential pillars in the treatment of choanal atresia. “It is by acts and not ideas that patients live”.

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